Agenesis of the dorsal pancreas associated with mucinous cysts and chronic calcific non-alcoholic pancreatitis

Dear Editor,

Dorsal pancreatic agenesis (DPA) is a congenital malformation which has been described in 107 cases (1,2). Here we present the case of DPA associated with chronic calcific non-alcoholic pancreatitis and mucinous cysts in the residual pancreas.

Case report

A 40-year old male presented with a 20-year history of chronic idiopathic cholestasis, hypercholesterolemia and type 2 diabetes mellitus. When undergoing endoscopic ultrasound, three cystic lesions of 13, 10.5 and 9 mm in diameter with multiple calcifications were observed in the head of the pancreas. Fine needle aspiration biopsy was performed and confirmed the mucinous content and a carcinoembryonic antigen (CEA) value of 2,335 ng/ml and CA-19.9 value of 197,455 U/ml. As a result, the patient was referred for surgery.

Computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) confirmed the three cystic lesions with calcifications and the absence of the neck, body, and tail of the pancreas (Fig. 1). Total laparoscopic pylorus-preserving pancreatoduodenectomy was performed. Macroscopic analysis revealed the head of the pancreas with multiple cystic formations with calcifications.

Fig. 1. Axial CT and T2 weighted MR images showing the pancreatic head with calcifications; there is no evidence of pancreatic tissue or duct in the expected position of the body-tail.

Fig. 2. Intraoperative ultrasonography showing a cyst (14 mm diameter) and a contiguous calcification (echogenic foci).
Microscopic analysis showed atrophy of the exocrine pancreas and cystic cavities formed by transitional epithelium without cellular atypia. We also observed amyloid deposits on the endocrine tissue.

**Discussion**

Since it was first described in 1911 (3), 107 cases of DPA have been reported (1,2). Its diagnosis requires confirmation of the absence of the neck, body, and tail of the pancreas and the duct of Wirsung.

Clinical presentation is highly variable, ranging from incidental discovery through to the development of pancreatic ductal adenocarcinoma. Diabetes together with abdominal pain are the most frequent signs. Recently, an association between DPA and chronic calcific pancreatitis and pancreatic adenocarcinoma has been reported (4). Other studies have reported a link with ampullary adenocarcinoma and intraductal papillary mucinous neoplasm (5).

**References**


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